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Anesthetic management and role in a patient with severe pulmonary hypertension with Eisenmenger physiology undergoing a cesarean section

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ABSTRACT

Objective: To elucidate the challenges associated with anesthesia and successful management strategies in a pregnant patient with severe pulmonary arterial hypertension and Eisenmenger physiology undergoing cesarean section. *Background:* Congenital heart disease is a leading cause of maternal cardiac deaths, with Eisenmenger's syndrome being high-risk due to potential complications. Pulmonary hypertension, characterized by an increase in mean pulmonary artery pressure, is associated with a high mortality rate between 30% and 56%. Advancements in medicine have improved the prognosis and quality of life for patients with pulmonary arterial hypertension. However, managing cesarean sections for parturients with Eisenmenger's syndrome remains challenging. *Case notes:* A 27-year-old primigravida with severe pulmonary arterial hypertension secondary to congenital heart disease, diagnosed with Eisenmenger physiology, underwent an emergency cesarean section at 32 weeks for optimal maternal and fetal outcomes. The multidisciplinary team opted for careful pre-operative assessment and invasive monitoring along with epidural anesthesia. Postoperatively, the patient faced complications, including thrombocytopenia, hematoma, and a positive COVID-19 diagnosis, yet was discharged in stable condition. *Conclusion:* Continuous vigilance throughout the perioperative and postoperative phases is crucial for optimizing outcomes in pregnant patients with Eisenmenger physiology. The successful outcome in this high-risk case underscores the importance of advanced medical modalities and multidisciplinary approaches in addressing the complexities of such pregnancies.

Keywords: Eisenmenger syndrome, pulmonary hypertension, cesarean section, high-risk pregnancy.

1. INTRODUCTION

Parturient experience significant physiological changes in several organ systems during a typical pregnancy. Reduced systemic vascular resistance (SVR), increased blood volume, and increased cardiac output (CO) as a result of elevated heart rate (HR) and stroke volume (SV) are among the modifications to the cardiovascular system. One of the leading causes of death for mothers is cardiac illness; in the past 30 years, congenital heart disease has been responsible for about 25% of maternal cardiac deaths (Gurumurthy et al., 2012). Eisenmenger's syndrome (ES) is categorized as high-risk due to the possibility of serious cardiac and newborn complications (Daliento et al., 1998). The phrase "Eisenmenger complex", which encompassed pulmonary hypertension and a large ventricular septal defect (VSD), was first used by Victor Eisenmenger in 1897. In 1958, Wood reinterpreted this as pulmonary hypertension linked to a patent ductus arteriosus or a reversed or bidirectional shunt, along with septal defects (Gurumurthy et al., 2012; Gupta et al., 2011; Chaudhari et al., 2023).

An increase in mean pulmonary artery pressure (mPAP) >25 mmHg at rest, as determined by right heart catheterization (RHC), is referred to as pulmonary hypertension. Pregnancy in women with pulmonary hypertension is known to be associated with a significantly high mortality rate between 30% and 56%, despite advancements in medicine (Weiss and Hess, 2000). About ten years ago, new advanced therapies for the treatment of pulmonary arterial hypertension (PAH) were developed, which has improved the patients' prognosis and overall quality of life (Macchia et al., 2007). Furthermore, and perhaps most significantly, the management of high-risk pregnancies has improved as a result of the adoption of a multidisciplinary approach, earlier identification of the underlying disease, enhanced comprehension of cardiopulmonary pathophysiology, and improved obstetric/anesthetic management (Stewart et al., 2001). Anesthetic management of cesarean section for parturients with ES continues to be a challenging task. This case highlights the intricate challenges and multidisciplinary approach required to manage a pregnant patient with severe pulmonary hypertension and Eisenmenger physiology undergoing cesarean section.

2. CASE PRESENTATION

A 27-year-old primigravida, diagnosed in 2019 with severe PAH secondary to congenital heart disease—specifically patent ductus arteriosus with a bidirectional shunt—exhibited features of Eisenmenger physiology, with an elevated mean pulmonary arterial pressure (PAP) of 66 mmHg additional to pulmonary vascular resistance (PVR) of 18.5 Wood Units (WU) on echocardiography. The management of her PAH involved a combined therapeutic approach with macitentan and sildenafil. The patient's obstetric admission on November 9, 2023, was prompted by concerns related to fetal surveillance. She presented with intrauterine growth restriction, manifested by an abdominal circumference (AC) at the 3rd percentile and absent end-diastolic flow (EDF) on Doppler imaging. Secondary polycythemia was evident with a hemoglobin (Hgb) level of 17.1 g/dl—a recognized complication of high-risk pregnancies in PAH patients. A multidisciplinary team consensus favored an elective cesarean section at 32 weeks of gestation to optimize both maternal and fetal outcomes.

Pre-operatively, the patient underwent a thorough cardiac and pulmonary assessment, with the active involvement of a cardiac anesthetist in perioperative care. During her hospital stay, continuous cardiotocography (CTG) monitoring revealed two unprovoked decelerations. With the unanimous agreement of the multidisciplinary team, an emergency cesarean section was deemed necessary, prompting the immediate transfer of the patient to the operating theater. Essential preparations included the availability of nitric oxide and extracorporeal membrane oxygenation (ECMO) in the operating room. Invasive lines, including central lines, arterial lines, and a pulmonary artery catheter, were placed for continuous hemodynamic monitoring. Epidural administration of 0.75% ropivacaine a total of 10 ml through the whole procedure. Prophylactic cefazolin was administered to prevent infection, and phenylephrine and norepinephrine infusions maintained hemodynamic stability, while on oxygen supplementation via a face mask at a rate of 4 L/min. The cesarean section proceeded without major intraoperative complications, but the premature delivery of a male baby weighing 720g necessitated admission to the Neonatal Intensive Care Unit.

Postoperatively, the patient was transferred to the Cardiac Surgery Intensive Care Unit (CSICU) for close observation. Her postoperative assessment revealed stability in central nervous system, cardiovascular, respiratory, gastrointestinal, and renal parameters. Hematological parameters showed a postoperative increase in Hgb to 20.4 g/dl, thrombocytopenia with a platelet count of 59 × 109/L, and a potential vitamin B12 deficiency, prompting the initiation of hydroxocobalamin treatment. A hematoma in the right

groin post-central line removal was identified, leading to ongoing oxygen therapy, deep vein thrombosis (DVT) prophylaxis, and antipulmonary hypertensive medications. Vascular consultation advised conservative management and platelet correction. Clexane was discontinued, and mechanical DVT prophylaxis was maintained. The patient's postoperative course was furthur complicated by a positive COVID-19 diagnosis on December 19. Despite these challenges, we discharged the patient from the hospital in a stable condition with regular out-patient follow up.

3. DISCUSSION

Maternal mortality in patients with ES was found to be correlated with the severity of pulmonary hypertension; however, a different study reported a range of 30–70% (Yuan, 2016). Guidelines for cardiovascular disease management during pregnancy state that pregnant women with simple congenital heart disease are either at no risk of maternal mortality after giving birth or have a slightly higher risk. However, because of intolerable hemodynamic changes, women with congenital heart disease and ES have a high mortality rate (Regitz-Zagrosek et al., 2018). Reduced right ventricular systolic function, ES, and severe pulmonary hypertension are associated with maternal mortality. According to a prior study, patients with ES and severe pulmonary hypertension had much higher mortality rates (Hartopo et al., 2019).

A fall in the SVR (which could permit a right to left intracardiac shunt) and thromboembolism (which could fatally interfere with an already embarrassed pulmonary circulation) are the two main issues that a pregnant patient with ES faces (Chaney, 1992). If oxygen therapy is not effective in treating fixed pulmonary hypertension, it could be a clear sign to end the pregnancy. The cardiologist recommended a scheduled cesarean section to prevent the stress of labor and the late stage of pregnancy. There are numerous reports regarding the use of elective cesarean sections to reduce risks to the mother at term and to optimize fetal outcomes in these patients (Makaryus et al., 2006). Given the complexity of our case, a multidisciplinary team meeting was organized leading to the agreement for an elective cesarean section at 32 weeks of gestation, aiming for optimal maternal and fetal outcomes.

However, there is a high risk associated with these patients receiving anesthesia. Maintaining cardiac output and SVR while avoiding hemodynamic alterations that could worsen the right-to-left shunt and raise PVR is the main objective of anesthesia in ES patient (Bennett et al., 2014). There are obvious hazards and drawbacks to general anesthesia. First, during laryngoscopy and intubation, catecholamine release may raise the PVR (Mishra et al., 2014). Secondly, under general anesthesia, positive-pressure ventilation reduces cardiac output and venous return. Third, pregnant women who undergo general anesthesia are more likely to experience reflux aspiration. Moreover, induction drugs decrease myocardial contractility and SVR. According to recent research by multiple authors, for ES patient with it is safe for epidural anesthesia to be administered (Pollack et al., 1990).

Catecholamine levels and PVR are lowered by epidural anesthesia through sympathetic blockade. Rapid changes in hemodynamics that exacerbate right-to-left shunting can be prevented by using epidural anesthesia, which acts more gradually than spinal anesthesia. Regional anesthesia was found to be safe and should be used in cases of Eisenmenger's syndrome, according to a review of 57 papers involving 103 patients (Martin et al., 2002). When epidural anesthesia is properly administered, hemodynamic and respiratory alterations are typically negligible. Both general and regional anesthesia have substantial morbidity and mortality, however, meta-analysis does not reveal a significant difference in perioperative mortality (Ammash et al., 1999). The epidural catheter also ensures postoperative analgesia and additional anesthesia. We consider epidural anesthesia as the optimal choice for these patients, aiming to maintain hemodynamics as normal as possible.

In pregnant women with ES, rapid hemodynamic changes may result in hypoxemia, right heart failure, pulmonary hypertension crises, and even unexpected death. Early detection of abrupt changes in hemodynamics is the aim of monitoring to start the right course of care and stop more problems (Anjum and Surani, 2021). On the other hand, the risk of infection and thromboembolism is increased by the placement of invasive monitoring equipment. The invasive monitoring role in ES is debatable, and as with any monitoring method, the complications risk must be considered. Patients with this condition often experience polycythemia, and the use of intra-arterial catheterization may be linked to an increased risk of thrombus formation following the procedure. A paradoxical air embolus and infection are possible risks associated with central venous catheter insertion (Ammash et al., 1999).

The challenges associated with pulmonary catheterization include the risk of pulmonary arterial rupture when pulmonary hypertension is present, in addition to the potential for arrhythmias and systemic embolization (Kandasamy et al., 2000). We inserted a central venous catheter, arterial catheter and a pulmonary arterial catheter, in this patient after weighing the benefits and risks.

Monitoring blood pressure invasively makes it easier to monitor real time hemodynamic changes. Additionally crucial to the decompensation of pulmonary hypertension is the postpartum phase, which is also when patients frequently experience sudden deaths. Following delivery, patients with ES need to be closely monitored at intensive care unit. Vigilant monitoring in conjunction with aggressive management is essential for a positive outcome. Pregnant patients with ES have the higher risk of developing thrombi, which can exacerbate pulmonary hypertension.8 Prophylactic antibiotic use against infective endocarditis should also be started (Diller et al., 2021).

Similarly, our patient was shifted to CSICU for close monitoring and maintained on oxygen, deep vein thrombosis (DVT) prophylaxis (40 mg of Clexane once daily), and antipulmonary hypertensive medication, along with 10 mg of Lasix twice daily. But she developed thrombocytopenia with a platelet count of 59 and a hematoma in the right groin post-central line removal. Clexane was held, and the patient was maintained on mechanical DVT prophylaxis. Complicating the clinical course, the patient also developed COVID-19. Despite these challenges, we discharged the patient from the hospital in a stable condition, emphasizing the successful multidisciplinary approach adopted throughout the perioperative period and the successful management of complex postoperative issues in this high-risk obstetric case.

4. CONCLUSION

In conclusion, a pregnant ES patient presents a significant challenge to medical professionals. The heightened mortality risk associated with this condition necessitates careful pre-operative planning and continuous vigilance throughout the perioperative and postoperative phases. For proper management of pulmonary hypertension, a thorough diagnostic evaluation with the goal of identifying multiple etiologies of the patient's current condition is necessary. It's important to fully comprehend the physiologic and hemodynamic changes associated with anesthesia as well as the available treatment options. A multidisciplinary team approach is also highly advised. Ongoing research and advancements in medical therapies, coupled with a collaborative and multidisciplinary approach, contribute to improving outcomes for pregnant patients with ES and similar high-risk conditions.

Ethical approval

Informed written consent was obtained from the patient

Author's contribution

Nasser Alabdulkarim, Nisar Soomro, Rahaf Althnayan: Participated in writing introduction, discussion and conclusion.
Salaheddin Zubek, Norah Almajed: Participated in writing introduction, case presentation and conclusion.
Alanood Alsaleem: Participated in writing introduction, discussion, abstract and conclusion.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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